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RHABDOMYOSARCOMA OF ORAL CAVITY

Dr. Kanupriya Gupta [1] | Dr. Jatin Gupta [2]

[1] Senior Research Fellow, Faculty of Dental Sciences, IMS, BHU, Varanasi (U.P.) INDIA-221005.

[2] M.D.S, Oral Medicine and Radiology, Varanasi (U.P.) INDIA-221005.

ABSTRACT

Rhabdomyosarcoma (RMS) is an aggressive malignant neoplasm of skeletal muscle origin that represents 50% of all soft tissue sarcomas in childhood, with most cases occurring in the head and neck. These are classified as parameningeal and non-parameningeal forms. These are the most common soft tissue sarcoma of the children, adolescents and young adults. Their etiopathogenesis and its molecular relevance have been emphasized. The first line of treatment is radical excision and this is usually supplemented by radiotherapy. It is believed that adjunct combination chemotherapy may greatly improve the prognosis. Inadequately treated tumors grow in an infiltrative manner and recur in a high percentage of cases. Bone does not constitute an effective barrier to the growth of the tumor and bone invasion is a frequent finding in head and neck rhabdomyosarcomas.

KEYWORDS: Rhabdomyosarcoma, oral diseases, diagnosis.

INTRODUCTION

Sarcomas are rare, malignant tumors that can arise from mesenchymal tissues at any body site. The histopathologic spectrum of sarcomas is broad, presumably because the embryonic mesenchymal cells from which they originate have the capacity to mature into striated skeletal and smooth muscle, adipose and fibrous tissue, bone, and cartilage(1).

Rhabdomyosarcoma was initially described by Weber in 1854 (2). The first published example of rhabdomyosarcoma, the malignancy of striated muscle, was probably a tongue lesion reported in 1854 (3). It is the most common soft tissue sarcoma of the children, adolescents and young adults. They are classified histologically into Embryonal, Botryoid, Alveolar and Pleomorphic varieties, and tend to occur predominantly in three regions: the head and neck, genitourinary tract, and upper and lower extremities (4).

Rhabdomyosarcoma may be defined as a malignant tumor of the rhabdomyoblasts with a microscopic picture simulating that of striated muscle cells (5). Although an uncommon lesion, this tumor is among the most common head and neck cancers in young persons (3).

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in pediatric patients and accounts for nearly 20% of soft-tissue sarcomas overall. In children, close to 50% of rhabdomyosarcomas arise in the head and neck, most commonly in the parameningeal region, orbit, oral cavity, nasopharynx, sinuses, ear, and neck (3).

CLINICALAPPEARANCE

The incidence of RMS is highest in children 1-4 years of age, falling to a lower rate at 10-14 years, and remaining steady between 15-19 years of age (6).It is rare after 45 years of age (3). A slight preference for males has been reported, with these tumors mainly occurring in the first and second decades of life (2).

The presenting signs and symptoms of RMS are variable, and are influenced by the site of origin, the age of the patient, and the presence or absence of distant metastases (1). The signs and symptoms may include pain, paresthesia, loss of teeth and trismus as a result of factors such as advanced tumor stage, infiltrative growth and tumor location (2).

A history of antecedent trauma is uncommon, and fever is only rarely present (1). The head and neck are the most frequently affected regions, followed by the orbit (35% of cases), trunk and extremities, intra-abdominal organs and genitourinary tract (23%) (2). Site predilection in the oral cavity varies according to different authors, some finding that the palate is the commonest site, while others report that the tongue is the most common site (6). There is general agreement that the orbit is the most commonly affected site. The nasopharynx and middle ear regions follow in frequency although at least one major study suggests that

the soft tissue of the neck may be the second most common site (7). The parameningeal sites, which include nasopharynx, nasal cavity, paranasal sinuses, pterygopalatine, and infratemporal fossa and middle ear, have been associated with extension directly to the central nervous system (CNS) in 35% of cases within 1 year of diagnosis (7).

The clinical appearance may exhibit smooth or lobulated surface, sometimes botryoid or grape cluster–like in appearance and definitely becomes fixed to surrounding tissues at an early stage (3).30% of all head and neck RMS have their origin in intraoral and pharyngeal structures. In affected patients, the tumor expands and infiltrates the muscle from which it arises, presenting first as a well demarcated nodule or polypoid lesion with a soft or gummy consistency. When these lesions grow rapidly they may cause dyspnea, dysphagia, and cough, including acute respiratory obstruction (8).

Initial symptoms may be vague, and may mimic other childhood and adolescent, soft-tissue sarcomas such as fibrosarcoma, leiomyosarcoma and neurofbrosarcoma (6). Of the sarcomas involving the oral cavity in children, rhabdomyosarcoma, alveolar soft-part sarcoma,fbrosarcoma, leiomyosarcoma, and Kaposi sarcoma are the most common (9).

However, neuroblastoma, another small cell tumor characterized by a diffuse pattern of small round cells and the presence of rosettes/pseudo rosettes with pale eosinophilic material is quite similar to the alveolar variant of rhabdomyosarcoma. The frequently elevated level of urinary catecholamines in neuroblastoma aids in the differential diagnosis (9-11).

The differential diagnosis also includes vascular malformations, within which the most common affecting the pediatric airway is the lymphatic or lymphatic-venous malformation (LM). Lymphatic malformations are typically described as cystic lesions present at birth that grow in proportion to the child. Although most head and neck LM are present at birth, they may not become clinically apparent until a child develops an upper respiratory infection or sustains trauma to the area (8)

TYPES

There are predominantly three types of Rhabdomyosarcomas:

1. **Embryonal variant:** Accounts for approximately 49%of all rhabdomyosarcomas and affects mostly children younger than 10 years of age, but it also occurs in adolescents and young adults. It is rare in patients older than 40 years of age.

2. **Alveolar variant:** It accounts for almost 30% of all rhabdomyosarcomas and tends to arise in patients of age group 10-25 years. It has predilection for deep soft tissues of the extremities. The tumor may arise at other places also though they are rare.

3. **Pleomorphic variant:** This is a rare variant which almost arises in adults older than 45 years. They arise mostly in the deep soft tissues of the extremities (4).

Enzinger and Weiss proposed two histogenetic possibilities for rhabdomyosarcoma: (a) primitive and undifferentiated mesenchyme origin and (b) embryonal muscular tissue origin (12-14).

TREATMENT

In the advanced stages of the disease, because of the infltrative growth, and depending on the site of the tumor, pain, paresthesia, loosening of the teeth, and trismus occur (6).Treatment therefore is by a multidisciplinary approach. It con-

sists of surgical removal of the tumor followed by multiagent chemotherapy with or without radiotherapy since RMS tend to metastasize to bone marrow, bone marrow aspiration should be a part of the staging procedure (4,5).

Oral rhabdomyosarcoma favourably is treated by radical surgical excision followed by multiagent chemotherapy,usually a combination of vincristine, dactinomycin, and cyclophosphamide. If complete resection is not possible then postoperative radiotherapy may be employed. Five-year survival rates have improved dramatically from less than 10% before the 1960s to 65% today. Stage I lesions have an even better prognosis (80%). Metastasis,when it occurs, is via either blood or lymphatic vessels, usually to cervical lymph nodes, lungs, bones, or brain(3).

PROGNOSTIC RELEVANCE

Oral rhabdomyosarcomas are classified within the nonparameningeal group of tumors, which do not tend to invade the central nervous system (6). As a result of their aggressive neoplastic behavior characterized by immature and highly invasive cells, RMS are associated with high rates of recurrence and generalized metastases through the hematogenic and/or lymphatic routes(2).

With the advent of combined surgical, chemotherapeutic, and radiotherapeutic management of RMS, the five-year survival rate is approximately 85% for this RMS subtype (6).

REFERENCES

1. Hicks J, Flaitz C. Rhabdomyosarcoma of the head and neck in children. Oral Oncol. 2002; 38:450-9.
2. Gordón-Núñez MA, Piva MR, Dos Anjos ED, Freitas RA. Orofacial Rhabdomyosarcoma: Report of a case and review of the literature. Med Oral Patol Oral Cir Buccal. 2008; 13:E765-9.
3. Gnepp DR. Diagnostic Surgical Pathology of the Head and Neck.W. B. Saunders Co.,Philadelphia, 2001. 550-1.
4. Batra R, Gupta DO, Sharma P, Bokariya P. Alveolar Rhabdomyosarcoma of Oral Cavity – A Rare Case. Al Ameen J Med Sci.2010; 3; 255-8.
5. Yueh-Lan H, Chin-Feng T, Li-King, Chen-Hua T. Rhabdomyosarcoma of the Adult Nasopharynx A Case Report. J Intern Med Taiwan. 2005;16:146-50
6. França CM, Caran EM, Alves MT, Barreto AD, Lopes NN. Rhabdomyosarcoma of the oral tissues – two new cases and literature review. Med Oral Patol Oral Cir Bucal. 2006;11:E136-40.
7. Peters E, Cohen M, Altini M, Murray J. Rhabdomyosarcoma of the oral and perioral region. Cancer. 1989;63:963-6.
8. Cirocco A, González F, Sáenz AM, Jiménez C, Sardi JR,Oscar RF.Embryonal Rhabdomyosarcoma of the Tongue. Pediatric Dermatology. 2005;22;218-21
9. Sekhar MS, Desai S, Kumar GS. Alveolar Rhabdomyosarcoma involving the Jaws: A Case Report. J Oral Maxillofac Surg. 2000; 58:1062-5.
10. Loducca SVL, Mantesso A. Intraosseous Rhabdomyosarcoma of the Mandible: A Case Report. Int J Surg Pathol. 2003;11:57-60.
11. Yoon SJ, Kang BC. Alveolar rhabdomyosarcoma involving the mandibular ramus and its surrounding tissues. Korean J Oral Maxillofac Radiol. 2004;34:111-6.
12. Gupta G, Budhwani KS, Ghritlaharey RK, Kushwaha A. Embryonal rhabdomyosarcoma tongue in a male child. J Indian Assoc Pediatr Surg. 2006;11:108-9.
13. Anik Y, Corapcioglu F, Aydin O. Rhabdomyosarcoma of the uvula.Pediatr Hematol Oncol. 2007;24:639-40.
14. Viswanatha B. Embryonal rhabdomyosarcoma of the temporal bone. ENT. 2007;4:218-22.